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CASE REPORT

Metastatic malignant gastrointestinal stromal tumor mimicking a right incarcerated inguinal hernia

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Summary Gastrointestinal stromal tumors (GISTs) are the most common malignant mesenchymal neoplasms of the digestive tract. Although surgery yields a high cure rate for low-risk GISTs, it can seldom cure high-risk tumors because the postoperative recurrence rate of GISTs is typically high; however, the prognosis substantially improves after imatinib mesylate therapy. A case is reported of recurrent GIST presenting as an incarcerated inguinal hernia to emphasize the importance of adjuvant therapy, particularly when treating advanced GISTs. Malignancy in the hernia sac should be considered if a patient has a history of intraabdominal malignancy.

1. Introduction

Gastrointestinal stromal tumours (GISTs) are common mesenchymal tumors arising in the wall of the gastrointestinal tract. Complete surgical resection is the primary treatment. However, postsurgical tumor recurrence is common, and adjuvant imatinib mesylate (IM) therapy is prescribed for patients with an increased risk of recurrence.[1] A patient who underwent surgical resection for a GIST over the distal jejunum is reported. Nevertheless, he was lost to regular follow-up after the wound stitches were removed and he missed IM therapy. Subsequently, he

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presented with a protruding mass over the right inguinal region and the pathological report confirmed a metastatic GIST.

2. Case report

The patient was an 82-year-old man who presented with a nonreducible, painful, and tender mass lesion over the right groin region. The patient, who was a retired porter with a history of chronic bronchitis, approached our general surgery outpatient clinic. Physical examination revealed a fixed, hard, painful, tender, and nonreducible mass over the right inguinal region. An inguinal hernia with incarceration was first suspected according to the physical examination. The patient's medical history showed that he had sustained a malignant GIST over the distal jejunum with rupture status following an operation within 2 years prior to this examination. However, we neglected to provide the patient with follow-up treatment at our outpatient clinic after the stitches had been removed. As the patient had a history of an intra-abdominal malignant tumor, a contrast-enhanced abdominal and pelvic computed tomography (CT) scan was immediately arranged because tumor recurrence was suspected, which may have herniated into the inguinal region. The CT results showed a well-defined heterogeneous mass lesion over the right inguinal region (Fig. 1). In addition, multiple intra-abdominal peritoneal carcinomatosis with lymphadenopathy in the pelvic region was noted. The patient presented with symptoms of persistent pain and signs of incarceration, and was at a high risk of tumor recurrence with herniation to the right inguinal region. The surgery indications were explained to the patient and arranged for surgical intervention after receiving his consent. When exploring the right groin, a sliding hernia was identified through the right internal ring with a stalk leading from the peritoneum (Fig. 2A). After opening the hernial sac, a well-capsuled, hard, ovoid mass with a dense adhesion to the hernial sac was identified (Fig. 2B and C). After dissecting the sac from the cord, high ligation was performed by removing the tumor with the sac simultaneously. The weak posterior wall was repaired using Bassini's technique. After opening the resected specimen, a 4-cm grayish-white tumor exhibiting no signs of necrosis or hemorrhage was observed. The pathology revealed a GIST composed of cigar-shaped interlacing cell fascicles and a high mitotic index (>5/50 HPF). Immunohistochemistry showed CD117 (+) (Fig. 3). Finally, the pathology report confirmed a recurrent metastatic GIST with herniation to the right inguinal region. Following surgery, imatinib (400 mg/d) was prescribed for the patient. After 3 months of treatment, contrast-enhanced abdominal and pelvic CT scans were ordered for the patient, and noted shrinkage of the lymphadenopathy over the pelvic region. No tumor recurrence or other complications were noted after a 2-year follow-up.

Figure 1 Ovoid heterogeneous mass over right inguinal region (arrow).

3. Discussion

GISTs are rare visceral sarcomas arising in the gastrointestinal tract wall, the muscularis mucosa, and muscularis propria, anywhere from the esophagus to the rectum. Its most common anatomic sites of origin are the stomach (60–70%), small intestine (20–30%), colon and rectum (5%), abdominal cavity (i.e., the peritoneum and omentum, 5%), esophagus (<5%), and retroperitoneal space (<3%).[2,3]

Surgery is the standard treatment for primary GISTs; however, surgical resection is seldom curative, particularly for large GISTs. Furthermore, >50% of the patients with GIST present with locally advanced, recurrent, or metastatic disease, making treatment difficult. The 5-year survival rate ranges from 50% to 65% following complete resection of a localized primary GIST, although this is reduced to ~35% for patients with advanced disease who undergo complete surgical resection.[4] The most commonly applied scheme for assessing the risk of recurrence is the consensus approach, which is based on the primary tumor diameter and mitotic count.[5] However, patients whose tumor has ruptured into the abdominal cavity, either before or during surgery, are at a high risk of tumor recurrence. This patient suffered from a GIST with a rupture, implying that the risk of tumor seeding was high. Furthermore, he had not received a regular follow-up at our outpatient clinic (including an abdominal CT scan and postoperative medication). Hence, we could not detect the recurrence earlier and take preventative measures until he presented at our clinic with symptoms mimicking an incarcerated inguinal hernia. Finally, an intraabdominal peritoneal carcinomatosis of the GIST with tumor herniation was identified in the right inguinal region.

Curative medication (IM) is available as a follow-up treatment of GISTs. Approximately 80% of GISTs have a mutated KIT gene, and 5% have a mutated alfa-type platelet-derived growth factor gene.[6] Imatinib is a potent, specific inhibitor of *KIT* exhibiting significant activity and tolerability in the treatment of malignant unresectable or metastatic GISTs. Previous research showed that imatinib either induced tumor shrinkage (≥50%) or stabilized the disease in a majority of patients.[5] Most

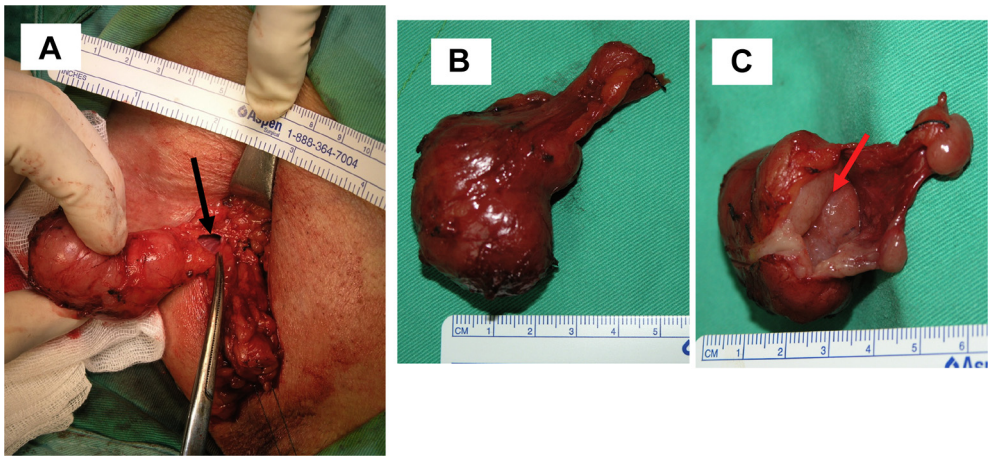

Figure 2 (A) Sliding hernia over internal ring with a stalk leading from peritoneum (arrow). (B, C) Transected hernia sac with a stalk. A grayish-white tumor without any necrosis or hemorrhage was noted after opening the sac (arrow).

patients (80—90%) with metastatic disease respond to imatinib or achieve durable tumor growth stabilization with continual therapy by using a daily dose of 400—600 mg.[5] Our negligence resulted in the loss of the patient to regular follow-up (abdominal CT), including the nonprescription of imatinib following the first surgery. However, the patient responded well to imatinib treatment following the second surgery, exhibiting shrinkage of an enlarged lymph node in the pelvis and no tumor recurrence. Therefore, we recommend that other physicians prescribe imatinib and conduct persistent follow-up treatments for the surgical intervention of GISTs, particularly for patients at a high risk of tumor recurrence, including those with ruptured or large tumors, or with a high mitotic index.

Incarceration occurs in approximately 5% of hernias. Emergency surgery is typically necessary because differentiating between viable and nonviable contents of the hernia sac is difficult. Previous studies have reported malignancies within the hernial sac when the hernia contains small pieces of the bowel and omentum. The first case of a tumor within a hernia sac was reported in 1749.[7] The most frequently reported tumors are of primary or metastatic colon cancer.[8] Any patient with a history of intraabdominal malignancy presenting with a new hernia should be examined for tumor recurrence. A history of intraabdominal neoplasms and systemic symptoms such as unexplained weight loss, anemia, altered bowel habit, or rectal bleeding indicates the possibility of a malignancy or colonic neoplasm.[9] If an abnormal nodular sac is noted during surgery, it should be examined to exclude a malignancy. The patient approached our outpatient clinic complaining of pain and tenderness. He presented with an irreducible mass lesion over the right inguinal region that mimicked the symptoms and signs of an incarcerated hernia. Had we been unaware of his previous case of the GIST, we could have misdiagnosed the patient's illness.

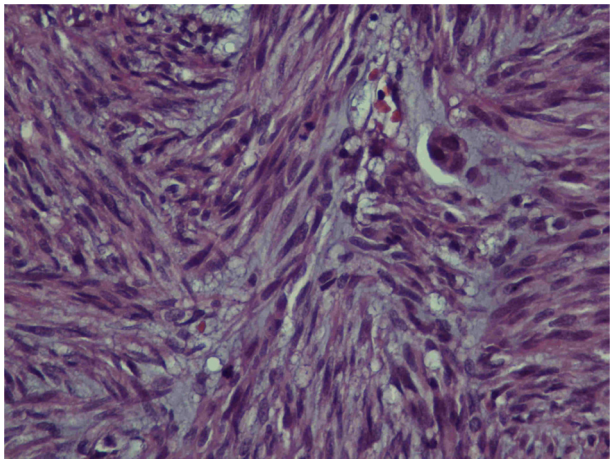

Figure 3 Stromal tumor present showing a spindled pattern with myxoid change and composed of cells arranged in distinct fascicles. The tumor is highly cellular with significant atypia (hematoxylin and eosin stain, × 400).

4. Conclusion

GISTs are common mesenchymal tumors arising in the wall of the gastrointestinal tract, typically reoccurring following surgery. The primary prognostic factors are mitotic activity and tumor size; furthermore, tumor rupture is a risk factor for postoperative recurrence. Thus, adjuvant IM therapy should be prescribed to patients at high risk of recurrence. In addition, the simultaneous occurrence of an inguinal hernia and cancer is rare, and easily overlooked. Relevant literature shows that the most common cause is either primary or metastatic colorectal cancer, other intra-abdominal malignancies have seldom been reported. In conclusion, patients with primary or metastatic GISTs can present with symptoms similar to those of incarcerated hernias. When diagnosing inguinal hernias, particularly among patients that exhibit a history of intraabdominal malignancies, physicians should consider whether malignancy is present. Contrast-enhanced abdominal and pelvic CT scans are effective tools for differentially diagnosing the case patient.

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